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Case Report

Pseudotumoral portal cavernoma: A rare case that challenges diagnosis ☆

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ABSTRACT

Portal cavernoma cholangiopathy (PCC), also known as portal biliopathy, refers to biliary duct abnormalities caused by extrahepatic portal vein obstruction (EHPVO) and subsequent cavernous transformation of the portal vein. Pseudotumoral portal cavernoma is a specific subtype of PCC characterized by the presence of numerous thin collateral veins that mimic the sheath of the common bile duct (CBD). We present a case of a 42-year-old women with pseudotumoral portal cavernoma secondary to portal vein thrombosis, a complication of myeloproliferative disorder. This case underscores the diagnostic challenges posed by pseudotumoral portal cavernoma and emphasizes the crucial role of imaging in achieving an accurate diagnosis.

Introduction

Portal cavernoma cholangiopathy (PCC) refers to anomalies of the biliary duct brought on by extrahepatic portal vein obstruction (EHPVO). Typically, this results in segmental stenosis of the common bile duct (CBD) due to compression of the peribiliary collateral veins. Pseudotumoral portal cavernoma is a specific variant of PCC characterized by the presence of numerous small collateral veins that mimic a mass surrounding the CBD [1].

A characteristic feature observed in patients with PCC is either moderately elevated bilirubin levels or the absence of symptoms.

Doppler color ultrasound (US), computed tomography (CT), and magnetic resonance imaging (MRI) are crucial for diagnosing PCC. Recent advancements in MRI and CT technologies, including helical scanning and multidetector computed tomography (MDCT), allow for rapid, artifact-free imaging. These developments facilitate comprehensive, noninvasive anatomical visualization of the portal venous and biliary systems [2].

We report a case of a 42-year-old woman diagnosed with myeloproliferative disorder who presented with elevated liver function tests. Our case study aims to elucidate the morphological changes in the portal and biliary tree in patients with pseudotumoral portal cavernoma, with the goal of reducing diagnostic errors.

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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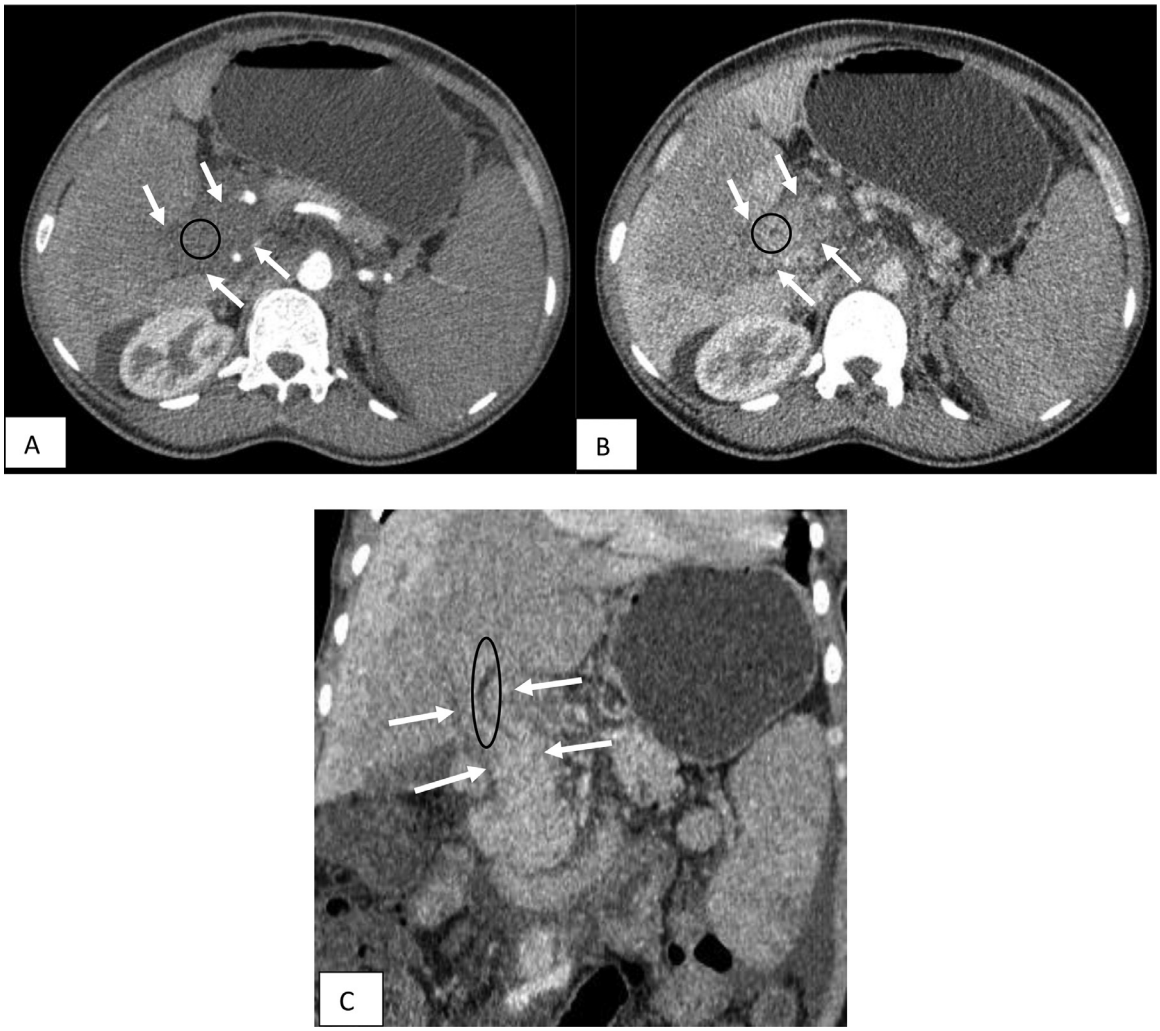

Fig. 1 – Axial CT scan of the abdomen in the arterial phase (A) and portal venous phase (B) coronal reconstruction CT scan (C) demonstrated a mass in the hepatic pedicle with homogeneous portal enhancement (arrow), causing compression on the CBD (circle).

Case report

A 42-year-old woman, with an unremarkable medical history, presented with asthenia lasting for 3 months. Physical examination showed hepatosplenomegaly but no other significant findings, including no fever or lymphadenopathy.

Laboratory results were as follows: Hemoglobin 16 g/dL(Normal value:12-15), Platelets 480,000/μL (Normal value: 150,000-450,000),Aspartate Aminotransferase (AST) 55 U/L (Normal value < 40), Alanine Aminotransferase (ALT) 60 U/L (Normal value < 50), Total bilirubin 2 mg/Dl (Normal value < 1.2), and Lactate Dehydrogenase (LDH) 300 U/L (Normal value, 140-280). Serum electrolytes were normal.

A bone marrow biopsy confirmed a diagnosis of myeloproliferative disorder. Due to elevated liver function tests, an abdominal ultrasound was performed, revealing hepatosplenomegaly and portal vein thrombosis (PVT).

For better characterization, an abdominal contrast-enhanced CT scan was performed. The CT scan demonstrated a tumor encircling the common bile duct CBD, causing its compression, The mass enhanced only during the portal phase. Hepatosplenomegaly and porto-systemic shunt, were noted. Additionally, a hepatic perfusion disorder was observed (Fig. 1).

Magnetic resonance cholangiopancreatography (MRCP) was performed, which demonstrated a sleeve in the hepatic pedicle developed around the CBD. It showed an intermediate T2 signal and enhanced homogeneously and intensely during the portal venous phase, but not during the arterial phase, causing compression of the CBD (Fig. 2).

The diagnosis of pseudotumoral portal cavernoma, a subtype of PCC was made.

The patient was placed on beta-blockers along with endoscopic variceal ligation for her PH. Regarding the pseudotumoral portal cavernoma, the decision was to monitor the patient closely and schedule a potential porto systemic shunt if there is biliary stenosis and/or jaundice.

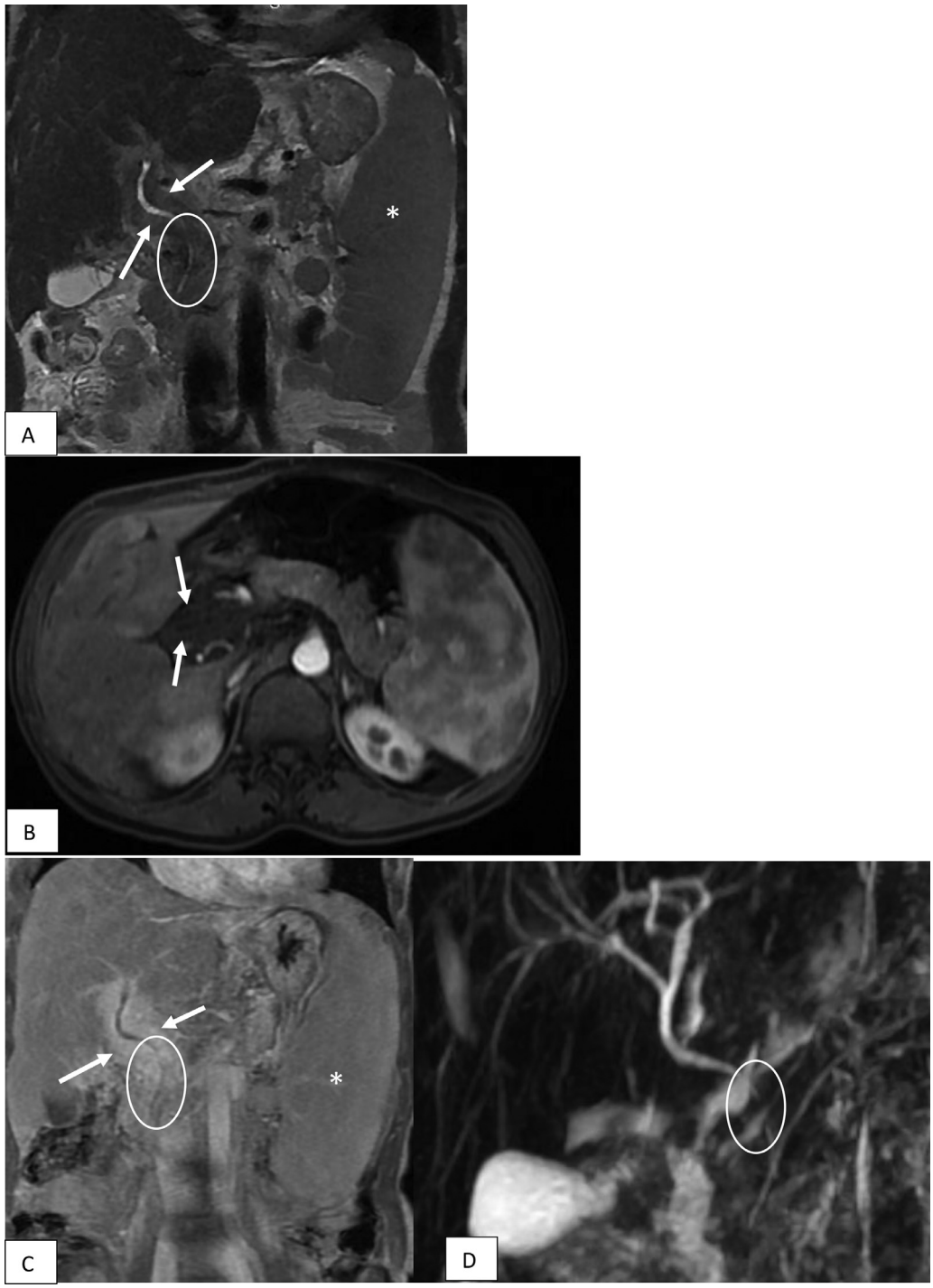

Fig. 2 – The coronal T2-weighted image (A) showed an intermediate signal sleeve developed around the CBD at the hepatic pedicle, this sleeve exhibited no enhancement in the arterial phase on Fat-sat T1 weighted image (B), with intense and homogeneous enhancement in the portal phase on Fat-sat T1-weighted image (C), causing compression on the CBD in the 3D-MRCP. Notice Significant splenomegaly was noted (*).

Discussion

PCC is characterized by abnormalities occurring at various locations within the biliary tree wall, typically resulting from extrahepatic portal hypertension (EHPVO). EHPVO most frequently develops due to extrahepatic portal vein obstruction (EHPVO). The underlying cause of EHPVO is often uncertain, and it may be influenced by a combination of local and systemic risk factors. In adults, EHPVO is commonly linked to conditions such as coagulation disorders, local inflammation, intra-abdominal infections, myeloprolif-

erative disorders, liver diseases, or hepato-bilio-pancreatic tumors [3]

The venous drainage of CBD is supported by 2 pericholedochal plexuses: the epicholedochal venous plexus of Saint and the paracholedochal plexus of Petren [3].

PCC can result from 2 mechanisms: either from external compression of the bile ducts by dilated portal collaterals or from ischemia caused by venous thrombosis [4].

There are 3 types of PCC: the fibrotic form, which is characterized by a predominant stenosis, usually located in the bile duct, and significant upstream dilation of the bile ducts; the varicoid form, which has multiple small stenoses with irregular contours; and the pseudotumoral form, which, as in our case, is characterized by numerous small collateral veins that mimic a mass surrounding the common bile duct [1].

Pseudotumoral portal cavernoma is typically asymptomatic despite associated biliary dilation; however, in certain instances, it may lead to the development of jaundice, cholangitis, or the formation of bile duct stones [5].

Color Doppler ultrasound (US) is the primary screening tool, typically revealing the absence of the portal vein and the presence of cavernoma formation [2].

CT imaging, employing thin slice sections and rapid acquisition times, provides high-resolution images of venous collaterals formed following portal vein occlusion and their impact on the biliary system, particularly through the use of portography techniques [6].

Currently, MRI has supplanted direct cholangiography as the preferred imaging modality for pseudotumoral portal cavernoma, with direct cholangiography now primarily used for interventional procedures. The recommended MRI sequences include routine T1-weighted and T2-weighted images, along with MR cholangiopancreatography (MRCP) and dynamic contrast-enhanced MRI (CEMR), also referred to as MR portography. These sequences enable simultaneous evaluation of biliary changes and portal collaterals [2,7].

The primary differential diagnosis for pseudotumoral portal cavernoma is hilar cholangiocarcinoma. Key diagnostic indicators include the absence of an enhancing mass lesion at the hilum with delayed contrast retention, the presence of PVT, and the visualization of dilated periportal collaterals around the area of biliary narrowing, which collectively support the diagnosis of pseudotumoral portal cavernoma [4].

Endoscopic ultrasonography (EUS) is useful for identifying CBD varices and bile duct stones. Furthermore, it helps differentiate between biliary strictures caused by pseudotumoral portal cavernoma and those resulting from cholangiocarcinoma or chronic pancreatitis [3].

Endoscopic retrograde cholangiopancreatography (ERCP) functions as both a diagnostic and therapeutic modality. It can reveal various abnormalities, such as ductal strictures, irregularities in duct caliber, segmental upstream dilation, ductal displacement, angulation or kinking, clustering of intrahepatic ducts, and ductal pruning [3]. Additionally, ERCP is used for therapeutic interventions, including stent placement and balloon dilation, to alleviate obstructive symptoms and improve bile flow.

Pseudotumoral portal cavernoma should not be biopsied as it can lead to catastrophic bleed and even death.

Treatment approaches for pseudotumoral portal cavernoma are customized based on patient symptoms. Asymptomatic patients with normal liver function tests generally do not require treatment. Initial management typically involves pharmacological therapy, with additional options including biliary stent placement and balloon dilation for cases that do not respond to medication. For symptomatic biliary obstruction that is not amenable to endoscopic intervention, a portosystemic shunt is recommended. Biliodigestive shunts are contraindicated unless the portal vein has been decompressed beforehand, due to the significant risk of bleeding [3,8,9].

Conclusion

In our case, the primary differential diagnosis was cholangiocarcinoma; however, visualization of PVT and dilated periportal collaterals adjacent to the site of biliary narrowing led to the diagnosis of pseudotumoral portal cavernoma. This diagnosis prevented unnecessary or potentially hazardous biopsy or therapeutic interventions, such as the risk of massive bleeding from hepatic pedicle dissection.

Patient consent

The authors of this manuscript declare that an informed consent for publication of this case was obtained from the patient.

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